Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION
44 EAST 23RD STREET, NEW YORK 10, N. Y.

Editor
BENEDICT F. MASSELL, M.D., Boston

Associate Editor
GORDON S. MYERS, M.D., Boston

VOL. XXIII FEBRUARY, 1954 No. 2

DISSECTING ANEURYSM OF THE AORTA*

At the present time the clinical diagnosis of dissecting aneurysm of the aorta is still made with considerable uncertainty, and all too few of these cases are diagnosed antemortem. With the rapid advances that are being made in vascular surgery, it is not inconceivable that methods may soon be devised for the repair of dissecting aneurysm of the aorta, and therefore it is important that physicians learn to diagnose this condition as promptly as possible.

It is encouraging to note that the various syndromes produced by dissecting aneurysm are gradually becoming clarified and that the correct premortem diagnosis is now being made with increasing frequency. Whereas only 6 of the 302 cases of dissecting aneurysm collected from the world's literature by Shennan[1] in 1937 were diagnosed antemortem, 25 per cent of our series of 44 cases reported in 1947 were so diagnosed[2]. In an additional 27 cases we have studied in the past five years, 44 per cent have been correctly diagnosed before death. A careful consideration of the clinical, laboratory, radiological, and pathological features of our 71 cases suggests that we can improve even further our clinical recognition of this usually fatal medical catastrophe.

Many reasons are offered to explain failure in making a correct diagnosis of dissecting aneurysm. Perhaps the most important is universal lack of suspicion. To quote Reich[3], "It is only through relentless correlation of the postmortem findings with the clinical picture in individual cases that a true clinical consciousness of the disease can be adequately established". Let us, therefore, consider some of the clinical features, laboratory features, radiological findings, and pathological aspects of this condition.

Clinical Features. Dissecting aneurysm of the aorta is twice as common in men as in women, roughly paralleling the sex incidence of aortic atherosclerosis. The great majority of cases occur in the fifth and sixth decades of life. However, the occurrence in younger people is not as rare as once believed. Many cases have been reported in individuals less than 40 years of age, particularly in association with pregnancy, coarctation of the aorta or other congenital cardiac disease.

The most striking single manifestation of dissecting aneurysm is the sudden occurrence of severe, excruciating, ripping or tearing pain. The pain will most often begin in the substernal area or in the upper abdomen. It may be followed promptly by stupor or coma. It may radiate to the neck, jaws, interscapular areas, back, abdomen, groin, testicles, hips or thighs. The radiation of the pain will depend on the point of beginning aortic dissection, the course of the dissection, and the organs involved. The time relations of the various types of pain may be of considerable help in making a clinical diagnosis. Failure of the pain to respond to large doses of narcotics should make one suspect dissection of the aorta. It must be remembered that in a number of cases, particularly those admitted in stupor, or coma, or shock, it will not have been possible to obtain a prior history of pain. The sudden occurrence of bizarre neurological disturbances is of especial significance. Dyspnea was also an important symptom in at least half of our cases, and other important symptoms were vomiting, confusion, disorientation, and collapse.

The physical findings will vary considerably, depending on the degree and sites of dissection, the age of the patient, the pre-existing cardiovascular disease, and the vessels and organs involved in the dissection. If collapse, shock, or coma do not supervene, the majority of patients will soon exhibit elevated temperature, tachycardia, and tachypnea. Signs of cardiac failure may develop. More than half the cases will have hypertension. This latter finding may be of especial value in helping to differentiate dissecting aneurysm from acute myocardial infarction. The absence of one or other radial or femoral pulse may be of considerable significance. Gouley[4] and others have emphasized a sign that may be almost pathognomonic of dissecting aneurysm, the development of the diastolic bruit of aortic

* From the Albert Einstein Medical Center and Philadelphia General Hospital, Philadelphia, Pennsylvania.

regurgitation. When sought for diligently, its discovery may enable one to make the diagnosis in suspected cases. Another rare but valuable diagnostic finding is the development of chest, abdominal, or lumbar ecchymosis in those cases having extravascular bleeding[5]. Other symptom complexes will be discussed further when the various clinical syndromes produced by dissecting aneurysm are considered.

Laboratory Features. Leucocytosis is an almost constant finding particularly if the patient survives the period of shock. Some of the white counts we obtained exceeded 25,000. Occasionally a rapidly progressive anemia may signify the collection of large volumes of blood in the wall of the aorta or the occurrence of extravascular hemorrhage. Many cases will exhibit elevation of the blood urea nitrogen. If the dissection involves the renal vessels, the urinalysis may reveal microscopic or even gross hematuria.

Radiological Findings. Most patients are too ill or do not survive long enough for satisfactory roentgen studies to be performed. Nevertheless, a number of cases have been reported in which there has been gradual widening or deformity of the aortic shadow thereby allowing the correct diagnosis to be made.

Pathological Aspects. Space does not permit a detailed consideration of the many pathological variations that have been described. There actually is no uniformity of opinion as to what precipitates dissection of the aortic wall. The first step, according to most observers, is the development of cystic degeneration of the media (Erdheim's medionecrosis cystica). A rupture of one of the diseased medial nutrient vessels produces a hematoma. This enlarges, the layers of the media are separated, and the intima is then split. The column of aortic blood then forces its way into the torn intima and produces various degrees of dissection along the medial coat. Dissection begins in the ascending aorta in over 70 per cent of the cases. In those cases in which medial necrosis is not found there is likely to be hypoplasia or coarctation of the aorta. This is particularly true in the cases of dissecting aneurysm occurring in patients under forty. The length of time that the patient survives will determine the other clinical features. Shock may be so severe that death is instantaneous. If the process is slower, the various bizarre clinical findings and syndromes occur. Death may result from rupture into the pleural, pericardial, or peritoneal cavities. Intractable cardiac failure may gradually develop over a number of days with eventual exitus. Slowly progressive retroperitoneal hemorrhage may occur, or the terminal picture may resemble uremia or saddle thrombosis of the aorta. An occasional case (as many as 10 per cent according to the late Soma Weiss) may recover. We have seen a case in which the new channel fashioned in the wall of the aorta by the dissection was the seat of extensive atherosclerotic changes.

Clinical Categories

In considering the various bizarre features of dissecting aneurysm, we have found it helpful to divide our cases into five clinical categories.

1. Cardiovascular. It is quite understandable that the most frequent and most difficult problem is to distinguish a dissecting aneurysm of the aorta from an acute myocardial infarction. Substernal pain, dyspnea, fever, leucocytosis, precardial friction rub, cardiac failure — findings typical of acute myocardial infarction — may all occur in dissecting aneurysm. The electrocardiogram may be abnormal but not diagnostic. If dissection involves the mouths of the coronary arteries, myocardial infarction may actually result. Making differentiation even more difficult is the fact that many individuals who have had one or more previous myocardial infarctions may present identical clinical pictures with the onset of the aortic dissection. In an occasional case the clinical findings may closely simulate primary iliac or femoral thrombosis and little attention will be given to the possibility of an underlying thoracic or abdominal aneurysm.

2. Cerebral or Neurological. The significance of cerebral or neurological abnormalities cannot be overemphasized. Thirteen of our 47 cases incorrectly diagnosed were thought to have suffered cerebral vascular accidents. Particularly is one apt to think of a cerebral accident if the patient is admitted in shock or coma without an antecedent history of pain. The development of bizarre neurological findings may be of considerable significance. Dissection may involve the coats of some of the spinal arteries and produce an "anterior spinal artery syndrome". In one of our cases the only symptom that led to a correct antemortem diagnosis was a sudden painless paraplegia.

3. Pulmonary. A third group of symptoms and findings may suggest primary pulmonary disease. A number of cases were thought to have pneumonitis or pulmonary infarction and

treated accordingly. The demonstration of hemothorax (usually on the left) should make one suspect dissecting aneurysm. Rupture of the aneurysm into the left pleural cavity is frequently a terminal event.

4. Abdominal. Many cases of dissecting aneurysm may masquerade as primary gastrointestinal disturbances. Chronic cases of dissection may erode into the oesophagus, stomach, or duodenum and simulate bleeding peptic ulcer[6]. In more than half of our cases pain began in the abdomen. The clinical picture may suggest peptic ulcer, acute pancreatitis, or mesenteric thrombosis. If an abdominal mass is felt, a diagnosis of gastric or colonic malignancy may be entertained.

5. Renal. In a small number of cases renal symptoms may dominate the picture. Pain in the back and hematuria will suggest renal calculus or infarction. Many of the cases become uremic as dissection involves the renal arteries or if shock itself produces secondary renal failure. In two of our cases the occurrence of back pain, hematuria, and lumbar ecchymosis led to the correct diagnosis of dissection of the aorta with retroperitoneal hemorrhage.

Summary

From this brief discussion it is apparent that dissecting aneurysm of the aorta may produce a number of clinical syndromes. A knowledge of these protean manifestations, and the realization that dissecting aneurysm may simulate primary cardiovascular, neurological, pulmonary, abdominal or renal disease, may help considerably in improving our diagnostic accuracy. It is our feeling that today we should make the correct diagnosis in more than half of our cases. This may be of more than academic importance. Although medical treatment at present is merely supportive, it is conceivable that future advances in vascular surgery may include repair of dissecting aneurysm of the aorta.

SAMUEL BAER, M.D.
5123 N. Broad Street
Philadelphia 41, Pennsylvania

The opinions and conclusions expressed herein are those of the author and do not necessarily represent the official views of the Scientific Council of the American Heart Association.

REFERENCES

1. Shennan, T.: Dissecting Aneurysm of the Aorta. Medical Research Council, London, 1939.
2. Baer, Samuel and Goldburgh, Harold L.: Dissecting Aneurysm. Am. Heart Jour. 35: 198, Feb., 1948.
3. Reich, N. E.: Dissecting Aneurysm of the Aorta. Clinics. 3: 349, 1941.
4. Gouley, B. A. and Anderson, E.: Chronic Dissecting Aneurysm of the Aorta. Ann. Int. Med. 14: 978, 1940.
5. Goldburgh, Harold L.: Personal Communication
6. Baer, Samuel and Loewenberg, S. A.: Aortic Aneurysms Simulating Disease of Gastro-intestinal Tract. Gastroenterology. 10: 617, April, 1948.

ANNUAL MEETING OF THE AHA, 1954

The Annual Meeting of the American Heart Association in 1954 will be held at the Conrad Hilton Hotel in **Chicago.** The Assembly Panels and the General Assembly will be held on Thursday and Friday, April 1 and 2, and will be followed by a specific scientific program on clinical cardiology on Saturday and Sunday, April 3 and 4, conducted under the auspices of the newly formed Section on Clinical Cardiology of the Scientific Council.

ANNUAL SCIENTIFIC SESSIONS, 1954, and THE SECOND WORLD CONGRESS OF CARDIOLOGY

The Second World Congress of Cardiology and the 27th Scientific Sessions of the American Heart Association will be held jointly in Washington, D. C., September 12-17, 1954. Membership in the Congress will be open to all members of the American Heart Association; nonmembers may attend by special invitation of the Organization Committee.

American and Canadian scientists desiring to present papers should submit titles with abstracts of not over 200 words, typewritten, double-spaced, in duplicate. They should be addressed to Charles D. Marple, M.D., Medical Director, American Heart Association, 44 East 23rd Street, New York 10, N. Y., and *must be received on or before April 1, 1954.*

The Formal Announcement of the combined meeting is being distributed widely to the medical profession. For further information, address Dr. L. W. Gorham, Secretary-General, Second World Congress of Cardiology, 44 East 23rd Street, New York 10, N. Y.